

## CASE REPORT

# Long term follow up of long QT syndrome treated by overdrive pacing

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**Abstract**

**Long term follow up of a patient with idiopathic long QT syndrome is described. A 5 year old girl was admitted with attacks of unconsciousness. Epilepsy was diagnosed and the patient was treated with anticonvulsants. During other episodes, ECG study showed torsades de pointes. The patient was treated with  $\beta$  blockers, stellatectomy without success, and later with overdrive pacing. The young woman is now 43 years old and in good health. It is suggested that early overdrive pacing be implanted in young people with symptomatic long QT syndrome.**

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Keywords: long QT syndrome;  $\beta$  blockers; pacemaker; long term survival

Two entities of idiopathic long QT syndrome have been described: the first by Jervell and Lange-Nielsen<sup>1</sup> in 1957; the second by Romano and others,<sup>2</sup> and later by Ward<sup>3</sup> in 1963. With the development of biomolecular technology, these syndromes were associated with chromosomal alterations.<sup>4</sup> Five genes were identified with more than 180 mutations. Not all long QT syndromes were associated with these alterations probably because other genes or mutations are implicated and not yet known. The mechanism by which the mutated gene leads to torsades de pointes or ventricular fibrillation is not well understood. Potassium and sodium channels are involved in prolonged action potential during which electrical dispersion may be the substrate of new depolarisation. This corresponds on the ECG to a

prolonged QT interval. To reduce this interval, several drugs were recommended. Agents still commonly used are  $\beta$  blockers, which increase background outward current and, in high concentrations, significantly depress the membrane stabilising membrane.

Recently, a combination of  $\beta$  blocker and pacing therapy has been proposed for the treatment of patients with drug resistance.

We report our long term experience with this combined treatment in a 43 old woman with an idiopathic long QT syndrome.

**Case report**

In 1974 we described a patient with Romano-Ward syndrome who had been followed up cardiologically since she was 14 years old. She had been previously hospitalised for seizure disorder with attacks of unconsciousness and treated with anticonvulsants since she was 5 years old. When she was 14 years old, during two consecutive menstrual periods she had two violent episodes of convulsions.

Her ECG during seizure crisis showed repeated spells of torsades de pointes and at rest she had a prolonged QT interval of 740 ms with a large T wave (figs 1 and 2).

Antiepileptic treatment was stopped. For her ventricular arrhythmia, she was initially treated with lidocaine and then with propranolol and digoxin.<sup>5</sup> Because of relapsing torsades de pointes, according to Schwartz and colleagues,<sup>6</sup> the patient had an extensive stellatectomy.

In 1978, the young woman presented with new episodes of torsades de pointes for which a pacemaker was implanted. It was a VVI pacemaker programmed at 100 beats/min. To prevent myocardial fatigue, in 1985 the pacemaker was reprogrammed at 90 beats/min. Despite combined treatment with  $\beta$  blockers (maximal tolerated dose) and pacing, she had other documented torsades de pointes, related to suboptimal pacemaker programming. In fact, ventricular pacing at rapid rates shortens the QT interval, eliminates pauses that precipitate torsades de pointes, and prevents further bursts of arrhythmias. Later a dual chamber pacemaker was implanted and reprogrammed in mode AAI at 100 beats/min (fig 3).

The follow up of our patient was marked by two recurrent episodes of ventricular arrhythmias caused by displaced lead or rupture. Our patient, now 43 years old, is still alive with a pacemaker implanted in 1978 and  $\beta$  blocker

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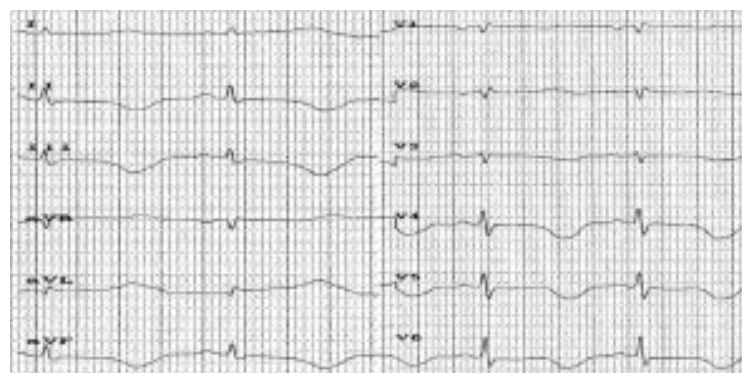


Figure 1 ECG at rest with a QT interval at 620 ms.

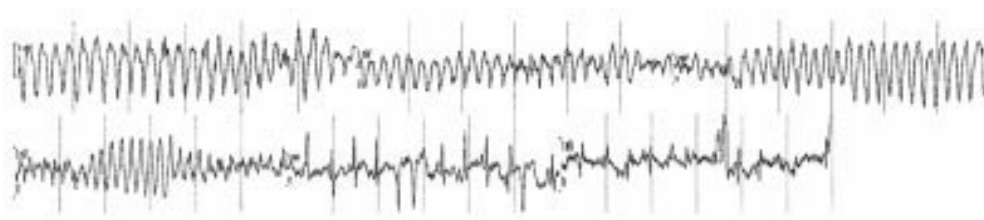


Figure 2 Continuous trace recorded during symptoms showing torsades de pointes.

treatment. Her left ventricular function has been monitored by cardiac ultrasound and is normal.

### Discussion

Patients with symptomatic idiopathic long QT syndrome have erroneously been considered to be “epileptic” patients and treated with anti-convulsants.<sup>7</sup> Our patient was initially suspected of having epilepsy but her ECG showed a classic torsades de pointes.

Few patients survive beyond the age of 30.<sup>8,9</sup> The long follow up and survival of our patient is a good example of improvement in treatment. The pacemaker prevents pauses that unleash ventricular arrhythmias. Its efficacy depends on a basic stimulation rate that shortens the QT interval.<sup>10</sup> Long term overdrive pacing did not lead to left ventricular dysfunction, which can induce new episodes of torsades de pointes. Lead malfunction can precipitate torsades de pointes. We have not observed other complications.<sup>11</sup>

We conclude: (1) that the incidence of idiopathic long QT syndrome is probably underestimated because of non-systematic ECG in young people; (2) that ECG is necessary in all paediatric patients who present with multiple seizure disorders and syncopal episodes;

(3) that perhaps some seizure disorders are associated with ventricular arrhythmias by the same mechanism (chromosomal mutation that lead to ion channel alteration); (4) that early overdrive pacing implantation associated with  $\beta$  blocker treatment may prevent death and aborted sudden death in idiopathic long QT syndrome; (5) that the overdrive pacing must be programmed at rapid rates to prevent pauses that precipitate torsades de pointes; and (6) that long term overdrive pacing stimulation does not impair left ventricular function.

We propose the use of early overdrive pacing implantation in association with  $\beta$  blockers. The AAI pacemaker is more physiological. A rapid rate is necessary to prevent pauses that precipitate ventricular arrhythmia. If torsades de pointes recurs, then a defibrillator must be implanted. In our patient, a simple defibrillator would not be appropriate because of recurrent, symptomatic ventricular arrhythmia, which would induce persistent discharge.

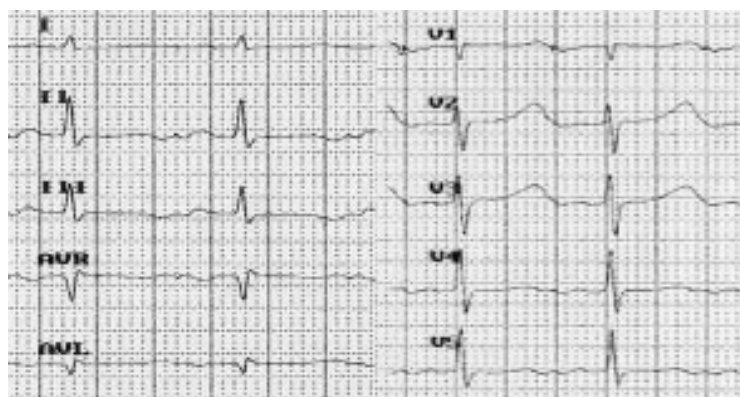


Figure 3 Pacemaker programmed in AAI mode at 100 beats/min. The QT interval shortens at 400 ms.

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